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Encapsulating Peritoneal Sclerosis

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1. Introduction

Chronic peritoneal dialysis (PD) can be complicated by encapsulating peritoneal sclerosis (EPS), a rare but the most severe complication associated with long-term PD. Morbidity and mortality are still high (range from 25% to 55%) especially in the first year after diagnosis. The international Society for Peritoneal Dialysis (ISPD) defined EPS by clinical signs of abdominal pain, bowel obstruction or weight loss in late stages of the disease. Clinical symptoms, radiologic findings and histologic criteria are the three diagnostic pillars.

During the course of the disease, development of adhesions causing symptoms of bowel obstruction often requires major surgery (figure 1A). Mostly, peritonectomy and enterolysis (PEEL) is the surgical treatment of choice.

Earlier stages of the disease are difficult to detect. Changes in transporter status or ultrafiltration failure can be first signs of EPS. The incidence of EPS increases with increasing time on PD, younger age, glucose load and peritonitis rate. EPS may occur when the patient is still on PD, but most patients become symptomatic after cessation of PD. In the minority of cases, EPS symptoms disappear and it seems to be a selflimiting condition. Actually, there exist no evidence based medical and surgical treatment options. Case reports and small case series are dealing with the effectiveness of immunosuppressants or antifibrotic drugs. But evidence for a specific medical treatment option is still lacking and prospective studies are needed.

2. Epidemiology of encapsulating peritoneal sclerosis

EPS is a very rare disease and the true incidence is still unknown. It is not exclusively seen in patients on PD but in this chapter we will focus only on PD-patients or former PD-patients.
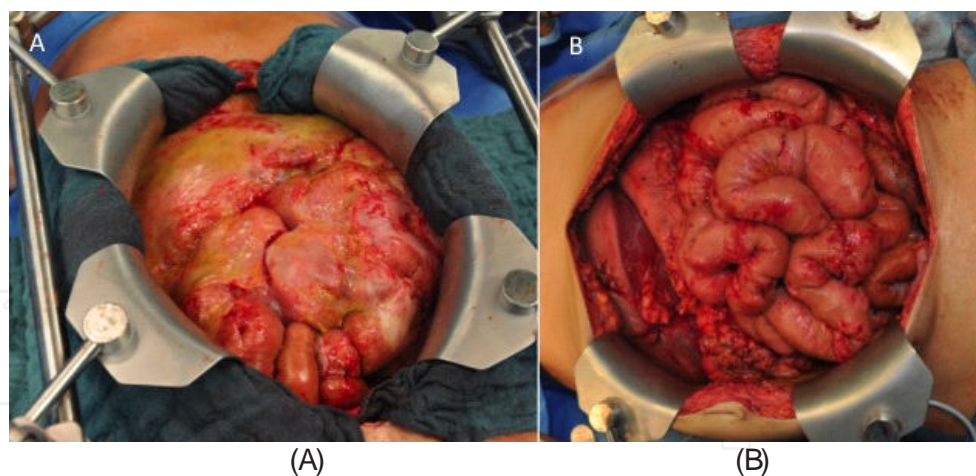
Figure 1. A 45-year old male presenting with massive abdominal pain, nausea, vomiting and weight loss over months. He was on PD for 72 months. EPS shows a sticky fibrin coating membrane on top of the bowel containing the brown and thick peritoneum B After an operation time of 420 minutes with peritonectomy and enterolysis (PEEL). Fibrin membranes were resected and restitution of intestinal function was achieved. 72 months after surgery, he has completely recovered and is back at work.

Kawanishi et al. reported 2004 and 2005 in their large cohorts, an overall incidence of 2.5% with an even higher incidence of up to 17.2% for patients on PD longer than 15 years. The Scottish Renal Registry included 1238 PD patients and Brown et al. showed that the incidence of EPS increases with time on PD: 2% after three to four years, 8.8% after five to six years and 5% after more than six years on PD. Interestingly, at the time of EPS diagnosis, 26% of the patients were still on PD whereas 72% were not on PD. Recently, Johnson et al. showed in a very large study from Australia and New Zealand a remarkably low incidence of 0.3, 0.8 and 3.9% after three, five and eight years on PD. In this study, the hazard ratio for patients receiving more than eight years PD was 12.1 in this study. It is noteworthy, that all existing data indicate that the majority of patients who are on PD for a long time will not develop EPS. The probability to develop e.g. endocarditis or osteomyelitis is much higher. Nevertheless, several risk factors for the development of EPS are discussed, but the published data are not uniform (table 1).

3. Diagnostic pillars in encapsulating peritoneal sclerosis

3.1. Clinical features of encapsulating peritoneal sclerosis

The clinical features of EPS are the result of acute or subacute small bowel obstruction mostly caused by adhesions and signs of systemic inflammation. The clinical findings mostly seen in patients with EPS are summarized in Table 2. In one of our EPS studies, all patients in the severe group (which was defined by the requirement for surgery due to extensive symptoms caused by bowel obstruction) had massive abdominal pain or vomiting. A large proportion in this group had both symptoms. Additionally, weight loss was noted in almost all patients in this group. In earlier stages of the disease, loss of peritoneal ultrafiltration capacity with weight gain (9 out of 31 in our study), a lower residual renal function, a higher glucose and icodextrin

Longer duration of PD
High Peritonitis Rate (severity/staphylococcus aureus)
High Glucose Exposure
Ultrafiltration failure
High volume regimen
Absence of residual renal function
Inadequate dialysis (low Kt/V)
Younger age
Cessation of PD (post-transplant)
Acetate buffer
Chlorhexidine
Icodextrin use
Medication (ß-Blocker, calcineurin inhibitors)
Smoking status

Table 1. Possible risk factors for EPS (most confident risk factors are marked in bold)

exposure are common. High transporter status in the peritoneal equilibration test prior to development of EPS has also been described in many EPS patients. These membrane changes are, however, not indicative for EPS, because they are also commonly observed in patients on long-term PD who do not develop EPS. Japanese investigators made an approach to subcategorize EPS in four stages. The first stage means the so-called pre-EPS stage with ascites followed by an inflammatory stage. Third stage is a stage of encapsulating of the bowel and the final stage includes symptoms of bowel obstruction. Up to now, this staging is not widely accepted in the international PD community.

Symptoms and clinical findings in EPS patients	
Signs of bowel obstruction	Signs of Inflammation
• Appetite loss	• Fever
• Nausea and vomiting	• Ascites
• Abdominal pain	• General fatigue
• Abdominal fullness	• Weight loss
• Diarrhea	Peritoneal adhesions
• Anorexia	• Bloody effluent
• Weight loss	• Ascites
• Feeling of heaviness	• Abdominal pain
• Constipation	• Abdominal mass
• Absent bowel sounds	

Table 2. Common clinical findings in patients EPS

3.2. Radiological findings in encapsulating peritoneal sclerosis

A CT-scan is mandatory in all patients with suspected EPS, but it is noteworthy, that no single diagnostic feature on CT-scan exists and any of the mentioned features can be found in scans of PD-patients without EPS. There is rarely more than one feature and in the majority of cases of low severity. Therefore, current evidence does not support the use of CT scanning to screen for EPS. Table 3 summarizes the typical imaging features of EPS and figure 2 shows an example of a 59-year old male with late-stage EPS. In very rare cases, x-rays show massive calcification (figure 3). Other studies like ultrasound, MRI or abdominal X-ray are insufficiently sensitive, rarely typical features are found.
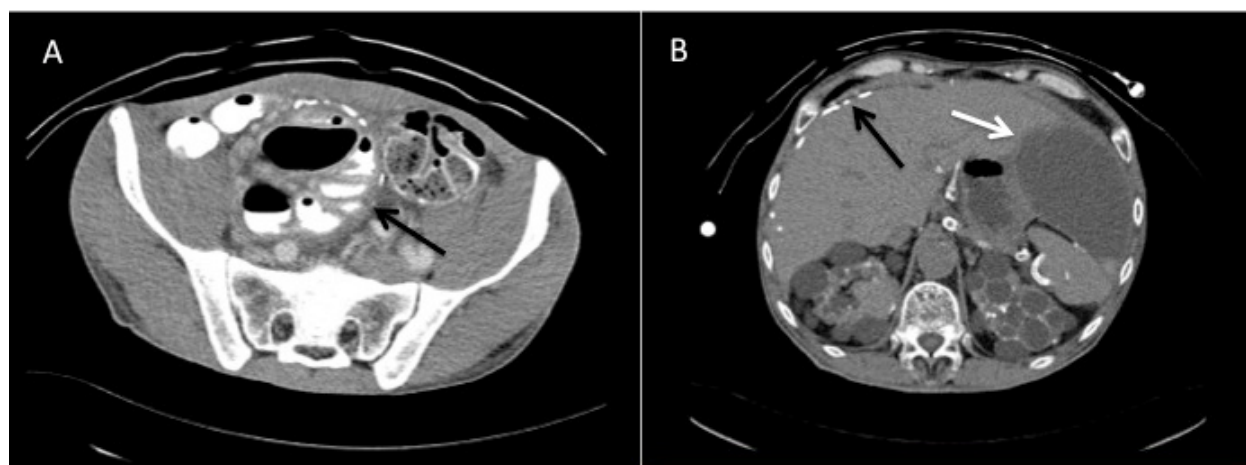
Figure 2. A CT scan showing typical „cocooning" with heavy calcifications and bowel obstruction (arrow). B Thickening, calcification and enhancement of the peritoneal membrane (black arrow). Loculated fluid collection (white arrow).

Figure 3. Peritoneal calcification (arrow) in a patient with established EPS after 22 years on PD.

Adhesion of bowel loops
Peritoneal thickening
Peritoneal calcification
Peritoneal enhancement
Bowel dilatation
Change of bowel calibre
Fluid loculation/septation
Thickening of the bowel wall

**Table 3.** Computed tomographic findings of EPS patients

3.3. Histological criteria for encapsulating peritoneal sclerosis

The third diagnostic pillar of EPS is based on the evaluation of peritoneal biopsies. The diagnosis of PD-associated pathologies, especially of EPS, is an interdisciplinary process, which requires, nephrologists and pathologists. The two most relevant pathologies of long-term PD are simple sclerosis (SS), which is a very common finding in PD- and EPS- patients. For a histological diagnosis, reproducible histological criteria are needed that can be used to differentiate the two entities. In 2003 and 2005 Honda and colleagues investigated peritoneal biopsies of 12 EPS patients. Fibrin deposition, fibroblast swelling, capillary angiogenesis and mononuclear cell infiltration were significantly more common in EPS than in peritonitis, ultrafiltration failure, uremia and so called "pre-EPS". Regarding the degree of these parameters, only fibroblast swelling and fibrin deposition exhibited were statistically significant different in their study. Several markers for fibroblast proliferation were also investigated. Garosi and colleagues investigated 224 peritoneal biopsies of non-EPS patients and compared the morphological findings with the biopsies of 39 patients with EPS. Significant findings in patients with EPS were thickening of the submesothelial cell layer, vasculopathy, arterial occlusion, inflammation, tissue calcification and ossification and arterial calcification and ossification (figure 4). In 2008 Sherif and colleagues compared peritoneal biopsies of 12 EPS patients with 23 non-EPS patients. Only fibrin deposition and the thickness of the compacta were significantly different between EPS patients and non-EPS patients. Actually, there is one main problem associated with most of the published data. In these previous studies, data acquisition was not standardized, observers were not blinded to the diagnosis and intra- and inter-observer variability was not given. Up to now there is no established method to differentiate between EPS and simple sclerosis. Especially in this field, further studies are needed to establish standardized histological criteria for EPS.

4. Pathogenic models of encapsulating peritoneal sclerosis

4.1. Epithelio-mesenchymal transformation (EMT) and the so-called two-hit-model

Over the years, the non-physiological properties of the PD fluids (glucose load, acidic pH, GDP`s affects the integrity of the peritoneal membrane. The insult of the serosa leads to
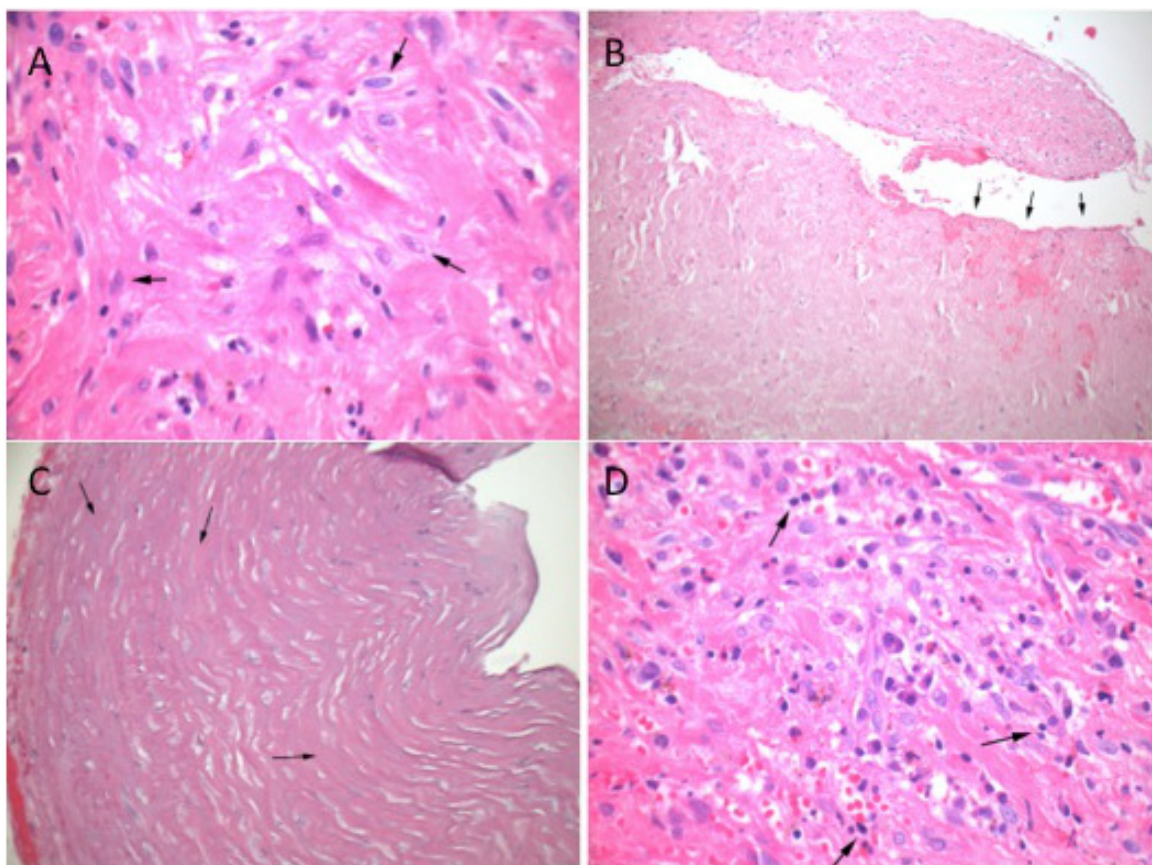
Figure 4. Peritoneal biopsies of EPS patients A HE staining showing an increased cellularity, round cells and fibroblast like cells (arrows). EPS, original magnification x400 B HE staining showing a decreased cellularity, fibrin deposits and a complete denudation of the mesothelial cell layer with fibrin exudations (arrows). EPS, original magnification x100 C HE staining showing a decreased cellularity with intracellular matrix (arrows), complete mesothelial denudation with fibrin exudations. EPS, original magnification x200 D HE staining showing fibroblast like cells, eosinophils, plasma cells and round cells (arrows). EPS, original magnification x400

secretion and production of different profibrinogenic mediators like transforming growth factor (TGF)-ß and of angiogenic factors like vascular endothelial growth factor (VEGF). The profibrinogenic factors lead to an increased fibrin deposition and neoangiogenesis. Additionally, the degradation of fibrin is reduced due to the loss of mesothelial cells and mast cells, which under normal circumstances produce fibrinolytic substances. This results in a so-called epithelio-mesenchymal transformation (EMT) (figure 5). As a consequence mesothelial cells change their function and become a more myofibroblast-like phenotype, which leads to the deposition of extracellular matrix and the promotion of fibrosis (figure 5). The described mechanisms lead to peritoneal fibrosis (PF), but do not necessarily proceed to EPS.

In the so-called two-hit-model of the pathogenesis of simple peritoneal fibrosis (PF) and EPS, it is postulated that PD itself is the first hit leading to the damage of the peritoneal membrane. When the second hit (e.g. an inflammatory stimulus (like a bacterial peritonitis)) occurs, EPS can develop. Others state, that EPS occurs in every patient on PD depending on the time on PD. Peritonitis rates, glucose load of the PD solutions and other factors might only influence this process.
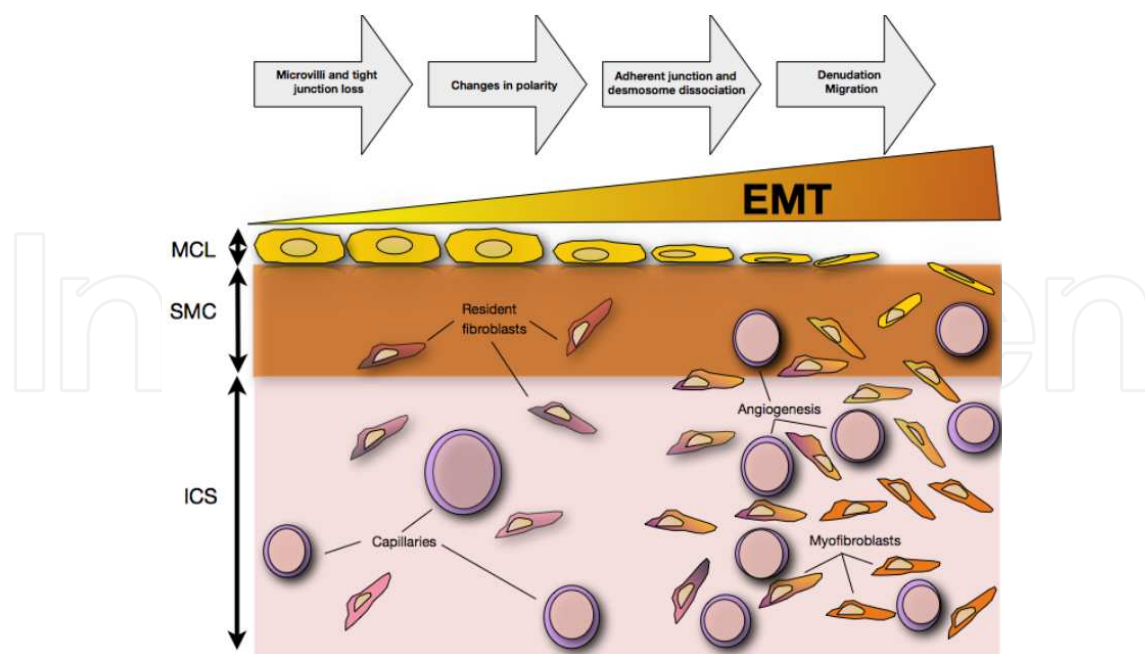
Figure 5. Mesothelial cells undergoing the so-called epithelio-mesenchymal transformation (EMT); MCL mesothelial cell layer; SMC sub mesothelial cells; ICS interstitial cell space. Adapted from Aroeira et al..

5. Management and outcome in encapsulating peritoneal sclerosis

The optimal management of EPS is not clear. Mortality and morbidity are still high (25% to 55%) especially in the first year after diagnosis. Table 4 shows a comparison of epidemiological studies of EPS patients. Up to now, there are no randomized controlled trials and the level of evidence is weak. The choice of surgical or conservative therapy is often based on the stage of the disease and varies quiet a lot between "EPS- centers". There is one prospective registry report from Kawanishi et al., who investigated 48 EPS patients in Japan. They report a recovery rate with total parenteral nutrition, corticosteroids and surgical treatment of 0%, 38.5% and 58.3%, respectively. All together, 37.5% of the patients in this study died, 45.8% of the patients recovered.

During work-up of patients with EPS, bacterial and fungal peritonitis must be ruled out before treatment might be considered. Treatment options include surgery and/or medical therapy.

5.1. Medical therapy

Steroids: Data about the use of steroids are not uniform, especially concerning dose and duration of therapy. Some studies suggested the administration of methylprednisolone pulse therapy with a dose of 500 – 1000 mg daily for 2-3 days, resulting in a reduction of inflammation and improvement of symptoms of bowel obstruction. Other groups recommend a dose of 0.5– 1mg prednisolone per kilogram of body weight daily for 2-4 weeks. In our referral- center we recommend an initial dose of 1 mg prednisolone per kilogram body weight for 4 weeks,

	Date of study	EPS cases	Study design	Mean PD duration (years)	Mortality rate (%)
Nomoto et al. (Japan 1996)	1980-1994	62	RS/MC	5.1	43.5
Rigby et al. (Australia 1998)	1980-1994	54	RS/MC	4.3	56
Lee et al. (Korea 2003)	1981-2002	31	RS/MC	5.8	25.8
Kawanishi et al. (Japan 2001)	1999-2001	17	PS/MC	10	35
Kawanishi et al. (Japan 2004)	1999-2003	48	PS/MC	4.3	37.5
Summers et al. (UK 2005)	1998-2003	27	RS/SC	6.1	29.6
Brown et al. (UK 2009)	2000-2007	46	RS/MC	5.4	56.5
Balasubramaniam et al. (UK 2009)	1997-2008	111	RS/MC	6.9	53
Johnson et al. (Australia 2010)	1995-2007	33	RS/MC	4.5	55
Kawanishi et al. (Japan 2011)	1993-2010	181	RS/MC	10.5	35.4
Latus et al. (Germany 2012)	1998-2011	42	RS/SC	6.5	21.4

Table 4. Comparison of epidemiological studies of EPS; RS, retrospective; PS, prospective; MC, multi-center; SC, single-center;

followed by a slow tapering over months depending on clinical symptoms and signs of inflammation. Especially in the so-called inflammatory period of the disease steroids may be useful. A prospective study demonstrated clinical improvement in 35.7% of cases treated with prednisolone. In patients with late-stage disease, histological analysis of peritoneal biopsies showed less acute or chronic inflammation. Therefore, the use of steroids is questionable.

Immunosuppression: The present evidence for the use of immunosuppressants (e.g. mycopheno-late or azathioprine) is mainly based on case reports. An increasing number of post-transplant EPS has been reported. One reason for the development of post-transplant EPS could be the widespread use of calcineurin inhibitors and the profibrotic potential of these drugs.

Antifibrotic agents: Several different antifibrotic agents are currently under investigation concerning development of peritoneal fibrosis and EPS. In animal models, inhibition of the renin-angiotensin-aldosterone system (RAAS) results in a decreased progression of peritoneal fibrosis. In induced EPS in rats, the blockage of RAAS resulted in a decrease of neoangiogenesis, peritoneal

thickening and ultrafiltration failure. Up to now, there exist no data regarding the use of angiotensin receptor blockers (ARB) or angiotensin converting enzyme (ACE) inhibitors in patients with EPS. But due to the low rate of adverse events and the widespread use of this medication in PD patients, inhibition of the RAAS should be the cornerstone of prevention of simple sclerosis and EPS. Tamoxifen, another antifibrotic drug, commonly used in the treatment of breast cancer, has been investigated in EPS patients. Tamoxifen revealed positive results in other fibrosing syndromes such as retroperitoneal fibrosis, fibrosing mediastinitis or desmoid tumors. Individual case reports and small case series supported the use of tamoxifen in EPS patients, mostly in combination with corticosteroids or as monotherapy. Recently, Korte and colleagues demonstrated in a retrospective analysis a survival advantage for patients with EPS treated with tamoxifen. Of the well-matched 63 patients with EPS, 24 were treated with tamoxifen and 39 were not. The mortality rate was significantly reduced in the tamoxifen group compared to the non-tamoxifen group (45.8% vs. 74.4%). The exact mechanism of action of tamoxifen in EPS is not understood. Some data suggest that an enhancement of transforming growth factor-ß (TGF-ß1) production stimulates metalloproteinase-9 to degrade type IV collagen. Other studies demonstrated an overexpression of TGF-ß1 which promoted fibrosis, peritoneal thickening and a loss of the capability of peritoneal repair. Therefore one mechanism of action of tamoxifen could be the inhibition of TGF. Other reports about the use of antifibrotic drugs like cholchicine or pirfenidone did not achieve acceptance in the PD community.

If medical therapy fails to improve the symptoms of EPS, surgical therapy must be considered.

5.2. Surgical therapy

Most data of operative treatment of EPS involve only small series or case reports. Macroscopically, late-stage EPS consists of two layers: a grossly thickened, leather-like peritoneum (EPS-membrane) and a white and opaque EPS-capsule covering the whole abdominal cavity. In contrast to the first description of the disease by Winne et al., EPS-capsule is the result of a dynamic process of shrinking. As a consequence, stricturing of the small bowel, sclerotic loop-to-loop adhesions and severe kinking of multiple bowel loops occur, causing symptoms of small bowel obstruction.

Although associated with a high morbidity and mortality rate, operative treatment probably represents the only realistic and potentially curative treatment for patients with late-stage disease. Because EPS is a rare disease, not all surgeons are familiar with the natural history of EPS and the required operative therapy. EPS is a disease of the visceral peritoneum and the serosa. Therefore, the operative treatment involves a complex procedure comprising *peritonectomy and intestinal enterolysis* (PEEL). Basic requirements of PEEL are the restitution of intestinal function and the prevention of recurrent disease. Simple adhesiolysis is not the treatment of choice. In fact, PEEL includes a demanding resection of EPS-capsule and EPS-membrane, whereas a partial resection of the small bowel serosa is unavoidable. Resection lines often involve the serosa or are located between the serosa and muscularis. With an incidence up to 20%, fistulas or anastomotic leaks are the leading complications after PEEL. Regarding this high morbidity rate, some authors suggest a protective stoma proximal to the first suture of bowel wall defects or anastomoses. Recently, we reviewed the treatment of 26 late-stage EPS patients at our referral

center regarding perioperative morbidity, mortality, and long term outcome. In our study, overall morbidity was 44% with minor complications in 2 patients (7%) and major complications in 11 patients (31%). Three patients (10%) died within the first year after operative treatment. These data suggest, that PEEL is a treatment option in patients with late-stage EPS that can be performed with acceptable morbidity (unpublished data).

Over a study period of two to 19 years, reported mortality rates vary from 25.5 to 56.6% especially in the first year after diagnosis (table 2). In our study including only late-stage EPS patients, three patients (10%) died within the first year after operative treatment, which is a favourable mortality rate (unpubslihed data). The overall mortality rate in our study with 42 EPS patients (31 of them required major surgery due to bowel obstruction) was 21.4%. To achieve such good outcome data, we believe, that these patients should be treated in specialized referral centers.

6. Conclusion

EPS is a rare complication of PD. There are three diagnostic pillars in EPS. clinical, radiological and histologic criteria. However a standardized approach is still lacking, histological analysis of peritoneal biopsies is important tool in the diagnosis of EPS. Peritoneal biopsies should be taken from all patients on PD at any time of surgery (e.g. catheter insertion, correction of a catheter malposition, catheter removal or any other abdominal surgery). Immunosuppressive therapy in patients with advanced disease might not be mandatory due to low degree of acute inflammation in these stages and the lack of prospective trials. Remarkably, time of first clinical symptoms consistent with to requirement of major surgery is very short. Therefore earlier diagnosis of the disease is mandatory, even in asymptomatic patients. Optimized operative therapy with PEEL represents a favourable treatment option in late stage EPS patients, which results in a low mortality and an acceptable morbidity rate.

Compared to the mortality rate of an age-matched dialysis population, outcome of patients even with severe EPS is not worse, if these patients are treated in specialized referral centers.

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